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Pterygium axillae as a rare manifestation of Poland syndrome



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ABSTRACT

Poland syndrome is characterized by a combination of absent pectoralis muscle, abnormalities of the rib cage, the breast, as well as brachy-syndactyly. We report a case of a 3 month old girl who was born with right sided axillary pterygium combined with flattening of the right anterior chest wall. Resection of a sclerotic band with reconstruction by Z-plasty was performed. Intraoperative and histopathological findings confirmed that the axillary pterygium developed on the basis of a scarred, hypoplastic pectoralis major muscle. Our case adds to the body of evidence that Poland syndrome may present in a heterogeneous fashion, including the rare finding of axillary pterygium with associated contracture. In these cases, early intervention prevents functional impairment.

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Poland syndrome is named after Sir Alfred Poland (1822–1872), who described a case in 1841 when he was a student-demonstrator in anatomy at Guy's Hospital in London [1].

It is typically characterized by absence of the costosternal portion of the pectoralis major muscle, absence or hypoplasia of the pectoralis minor muscle, abnormalities of the rib cage, hypoplasia or absence of the breast nipple and brachy-syndactyly [2]. In fact, there is a large variety of clinical presentations; but in most patients partial or complete absence of the pectoralis major muscle can be found [3]. Rarely, all characteristics are detected in one individual [2,4]. Despite the absence of the pectoralis muscle, there is seldom a significant functional muscle weakness in the typical case [2].

The defect is more often located on the right side than on the left side, bilateral occurrence is extremely rare [3]. The incidence has been estimated to occur in 1:30 000 newborns [5], with male predominance ranging from 2:1 to 3:1 [4]. On the affected side, the breast and the nipples, if present, are usually displaced slightly upward [3].

In such cases where surgical reconstruction is indicated, several different techniques have been described by thoracic and plastic surgeons, ranging from chest wall prostheses, mammary prostheses, latissimus dorsi muscle transfers, transverse rectus abdominis musculocutaneous flaps, sternal/rib reconstruction, or a combination of methods [6]. When stabilization of the chest wall as well as breast augmentation is needed, single-stage reconstruction is generally recommended to provide best cosmetic results [2].

1. Case report

A 3 month old girl presented to our department with a unilateral right-sided axillary pterygium combined with a flattening of the right anterior chest wall and functional impairment. The congenital web of the right axillae significantly restricted abduction of the arm beyond 80° (Fig. 1). In combination with the flattening of the right anterior chest wall as a clinical sign for the absence of the pectoralis major muscle, as well as relative hypoplasia of the ipsilateral nipple,



Fig. 1. Pterygium axillae severely limiting abduction and elevation of the right arm.

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Fig. 2. Release of the fibrous band and cutaneous Z-plasty provides an excellent functional and cosmetic result. Full abduction (left) and elevation (right) is easily achieved at follow-up 10 weeks after surgery. Note the absence of the pectoral musculature on the patient's right with the corresponding indentation inferior to the scar.

we made the diagnosis of Poland syndrome. Further clinical examination ruled out other features, such as, abnormalities of the rib cage or symbrachydactyly. In order to prevent further contracture of the right arm, and to allow normal motor development, we planned release of the underlying contracture with reconstruction by Z-plasty at the age of 9 months.

During the procedure, a fibrous band limiting the range of motion was found and excised down to deep fascia, and a cutaneous Z-plasty flap was created to release the axillary contracture. Intraoperatively, we identified a rudimentary part of the pectoralis major muscle in continuity with the excised fibrous strand inserting in the anterior chest. Histopathology confirmed the described myosclerotic findings. The postoperative course was uneventful and the girl was discharged home on postoperative day 2 with full range of movement of the right arm in reference to the left. At follow-up 10 weeks later, the patient maintained her full symmetric and unrestricted range of motion. The Z-plasty scar had healed well (Fig. 2).

2. Discussion

In this special case of Poland syndrome, the hypoplastic, sclerotic pectoralis major muscle resulted in an axillary pterygium. This is the first case of this kind of manifestation reported in literature. Two theories concerning the development of the syndrome have been proposed. One of them postulates a lethal mutation of an upper limb bud cell line which leads to hypoplasia or aplasia. The other, so called 'vascular hypothesis', suggests a regional vascular incident of the subclavian vessels which leads to muscle absence [7].

The vast majority of procedures described for Poland syndrome are performed after completion of growth in adolescent or adult patients [7]. Besides cosmetic problems, functional restrictions, and other rare symptoms such as lung herniation are often quoted as indications for surgical treatment [2]. Patients with Poland syndrome often experience body image disorders and decreased quality of life. A recently published study evaluated this question in regard to surgical timing. The authors showed that patients under the age of 20 had the highest scores for standardized "Body Uneasiness Tests". Interestingly, patients regained similar scores

compared to their peers in the control group after surgical correction. The article concludes that surgical planning should be started early in the period of growth [8].

In our case, the patient not only had a cosmetic impairment, but severe limitation of her right brachial range of motion. Developmental milestones in the first year of life strongly depend on normal upper limb mobility, such as grasping and transferring objects, self-feeding finger-foods, and pulling up to a stand. We therefore felt that early intervention in our patient was crucial to avoid motion impairment and subsequent potential developmental delay.

Regular follow up with a special focus on lung function, muscle function and breast development is planned. The parents were advised that when their daughter reaches puberty, she may require further surgical intervention including breast reconstruction or augmentation.

3. Conclusion

This case adds to the body of evidence that Poland syndrome may present in a heterogeneous fashion, including the rare finding of axillary pterygium with associated contracture. If treated early and aggressively, long-term sequelae in terms of functional impairment may be avoidable.

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